

pathologically to be related to a plaque in the lower medulla.

Although a diagnosis of multiple sclerosis in life was felt to be likely in our patient, the clinical presentation, the stepwise progression with no previous history of neurological dysfunction to a final, irreversible, locked-in state, was unusual. Furthermore, oligoclonal bands were absent from the CSF on two occasions, a finding usually seen in only 30% of patients with multiple sclerosis. Subsequent transverse myelitis of the spinal cord showed dissemination of the disease in time and place. Furthermore, the midbrain/brain stem demyelinating lesion showed the typical pathological features of multiple sclerosis. The asymmetry and extent of this lesion are features against a diagnosis of central pontine myelinolysis. Necrotising myelopathy of the severity seen in this patient may occur in association with carcinoma, but is probably more often associated with plaques of multiple sclerosis elsewhere in the nervous system.⁵ Overall, the diagnosis of multiple sclerosis best fits the clinical and pathological features of this most unusual case.

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Spontaneous ejaculation secondary to spinal cord disease

Spinal cord lesions in humans are associated with impaired ejaculation,¹⁻³ suggesting that the effects of descending pathways on the spinal ejaculatory mechanism are predominantly excitatory. In rodents, however, there may be an additional and important descending inhibitory pathway.⁴ In keeping with the presence of a similar inhibitory pathway in humans, we report the case of a patient in whom a spinal lesion was unusually associated for a period of time with spontaneous or easily provoked ejaculation.

The patient, a 70-year-old man, had noted an episode of spontaneous ejaculation when his glans touched the side of a catheter bottle during admission to hospital for a myocardial infarction five years previously. About every three weeks thereafter, until shortly before this report, he ejaculated at the onset of micturition, in association with symptoms of urinary urgency and the sensation of bladder fullness. Spontaneous ejaculation also occurred occasionally during midstream micturition. These episodes occurred with a flaccid penis and with a buildup of pelvic floor tension followed by minor, rhythmic contractions of the pelvic floor which he described as a "mini-orgasm".

He had not had any spontaneous erections since the onset of these symptoms, though sexual function had previously been normal.

Two years before presentation he developed constant lower limb paresthesiae sometimes affecting the buttocks. These sensory symptoms were unpleasant, resolved within five to 10 minutes of lying flat, and resumed as he placed his feet on the floor. For two years he had had worsening stiffness of his legs, which was aggravated by prolonged walking and sometimes associated with discomfort in his buttocks. In recent months he had noted spontaneous jerks of his legs when lying flat. He had been in atrial fibrillation since his myocardial infarction and he described a number of carotid transient ischaemic attacks without any abnormalities on the cranial CT scan or digital subtraction angiography of his neck vessels during subsequent investigation.

On examination his blood pressure was 150/90 and his pulse irregularly irregular. The dorsalis pedis pulses were absent but the rest of the peripheral vascular examination was normal. His cranial nerves and arms were also normal, but in his legs there was a moderate spastic paraparesis with hyperreflexia and extensor plantar responses. His abdominal reflexes were preserved and there was no sensory loss. Bulbo-cavernosus and anal reflexes were present, as well as the left cremasteric reflex.

Routine haematological and biochemical tests were normal and syphilis serology negative. The electrocardiogram confirmed atrial fibrillation. Urodynamic studies indicated the presence of detrusor hyperreflexia and marked detrusor-sphincter dyssynergia, with a residual bladder volume of 250 ml. Cerebral MRI, analysis of the cerebrospinal fluid, and a number of electrophysiological studies were normal, including lower limb nerve conduction studies, tibial and pudendal somatosensory evoked potentials, brainstem auditory and visual evoked potentials, and finally electromyography of the anal sphincter. A full myelogram and computed myelography a L4-5 and L5-S 1 were also normal. MRI of the thoracolumbar region (figure) showed a high signal lesion extending over the whole mid-thoracic cord. The lesion was situated in the anterior part of the cord and was bilateral. The cord was not expanded on the myelogram or in the MRI study. Overall, the results of the imaging studies suggested that the cord lesion was ischaemic in origin.

Although the patient's ejaculate was not tested for the presence of semen, the description of his symptoms favours the occurrence of true ejaculation rather than seminal emission or the passage of urinary gravel, as rhythmic contractions of the pelvic floor accompanied the expulsion of fluid. The most unusual feature in this patient was the symptom of such easily provoked ejaculation in the presence of a spinal cord lesion. Earlier reports of this association have been anecdotal,⁵ and cord pathology is generally found to lead instead to impairment of ejaculation. It is known, however, that a reflex mechanism exists in the T9-L2 (sympathetically driven emission of semen) and S2-3 cord segments (somatically driven propulsion of semen) from which involuntary ejaculation may be produced.^{2,3} Thus in patients with spinal injuries above T9, emission can be induced using electrovibration or sympathetic stimulation below the level of the lesion.^{2,6} In rodents this spinal sexual mechanism receives descending inhibition to pudendal motor neurons and lumbosacral inter-



T2-weighted MRI (SE 1500/80) showing a high signal lesion in the patient's mid-thoracic cord (A, arrow). In axial images the lesion occupied the anterior part of the cord (B, arrow). Images were obtained using a 0.5 T system (Picker International).

neurons, and in male animals with a spinal cord transection ejaculation can be induced by fluid distension of the urethra⁴ in a manner reminiscent of our patient's symptomatology. In humans the overriding supraspinal influence on sexual function is usually excitatory, but it is possible that spinal lesions could result in a situation resembling that in rodents by selectively interfering with an additional, smaller descending inhibitory pathway projecting to the spinal ejaculatory mechanism. Support for this argument may also be found in human spinal injury where a variety of stimuli, including bladder distension, can induce exaggerated autonomic responses such as sweating and hypertension.¹ As emission is primarily under sympathetic control^{2,3} it is at least theoretically possible that our patient's symptoms could be viewed as a form of autonomic disinhibition, in this instance affecting the whole reflex ejaculatory mechanism.

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Abnormal evoked potentials in Miller-Fisher syndrome: further evidence of combined peripheral and central demyelination

We read with interest the short report by Ferrer *et al.*¹ about a patient with Miller-Fisher syndrome in whom CNS demyelination was documented by MRI a few years later.¹

We would like to direct attention to an additional case supporting the possibility of combined central and peripheral demyelination in this syndrome.

A 66-year-old previously healthy woman was referred for diplopia of 24 hours duration, an unsteady gait, and distal paresthesias in the upper limbs. Examination showed bilateral ptosis, complete external ophthalmoplegia, and severe axial and limb ataxia. On the second day of admission to hospital, progressive proximal muscle weakness developed accompanied by generalised areflexia. The plantar responses were flexor. All sensory modalities were intact. Routine laboratory examinations, CT of her brain, and EEG were normal. The sterile acellular cerebrospinal fluid obtained on the fifth day of admission to hospital was clear and under normal opening pressure, with a protein concentration of 0.75 g/l and no oligoclonal bands.

Electrodiagnostic studies disclosed prolonged distal motor latencies, slow conduction, and prolonged F-responses in the median, ulnar, common peroneal, and posterior tibial nerves. Needle electromyography indicated active symmetrical generalised denervation. The visual evoked potentials (VEPs) were remarkable for prolongation of the p100 peaks (right 126 ms; left 128 ms; normal 100-110 ms), which retained their normal amplitude. Brain stem auditory evoked potentials (BAEPs) showed an impaired wave pattern with prolongation of N₃, N₅, and brain stem conduction time.

Within one week and with only conservative treatment, spontaneous improvement was noted in the ptosis and horizontal eye movements, but there was still partial limitation of vertical gaze.

Neurological examination after three months showed a full range of eye movements, normal muscle strength, and deep tendon reflexes. The VEPs and BAEPs had returned to normal.

The aetiology and pathogenesis of the

Miller-Fisher syndrome are not well outlined, particularly the location of the pathological changes. Most workers favour a peripheral origin,² although others suggest brain stem inflammatory lesions³ or a combination of central and peripheral demyelination.⁴ Abnormal BAEPs have been described in only a few cases.⁵ Results from MRI have usually been normal,⁶ except in the reports of Giroud *et al.*⁷ and Ferrer *et al.*¹

Abnormal BAEPs and VEPs, together with evidence of demyelination on brain MRI, support the hypothesis that in the Miller-Fisher syndrome there is a combination of peripheral and central demyelination. Considering the dissimilarities between central and peripheral myelin, however, this conclusion leaves the immunological mechanisms by which this process is mediated unclear.

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Transient sixth-nerve palsy as the first presentation of acute leukaemia

It is not unusual for the cranial nerve to be affected early in the course of acute lymphoblastic leukaemia (ALL) and this is considered to be the result of meningeal infiltration.^{1,2} Unless specifically treated, the neurological deficit is usually progressive. We report here the case of a patient with transient sixth-nerve palsy as a presentation of ALL.

A 43-year-old man was admitted with an acute onset of diplopia. He had a three month history of malaise and weight loss. He had previously had a systemic evaluation, which was normal except for a Westergren erythrocyte sedimentation rate of 40 mm/h, a transient disturbance in liver function tests (aspartate transaminase, alanine transaminase) and a constant increase in lactate dehydrogenase. Serological tests for hepatitis were negative.

On examination a complete left sixth-nerve palsy was found. The rest of the general and neurological examination was normal. A complete blood count and film, blood glucose concentration, thyroid function tests, antinuclear factor, serum

immunoglobulins, immunoelectrophoresis and antibodies to acetylcholine receptor were all within normal limits. Lumbar puncture yielded a clear acellular fluid with 530 mg/l protein and 3.6 mmol/l glucose. Computed tomography of his brain gave normal results. MRI of his brain showed homogenous replacement of the skull bone marrow by an enhancing material with extension to the left cerebello-pontine angle and left cavernous sinus. A bone marrow specimen was therefore obtained and showed extensive infiltration with immature lymphoid cells, compatible with ALL.

A week after admission a gradual, spontaneous improvement in eye abduction was noted. Several days later, before any treatment (and specifically without steroid treatment), his eye movements were full in all directions, and there was complete resolution of the diplopia. The patient was treated with a standard chemotherapy protocol for adult ALL. An Ommaya reservoir was inserted and intrathecal methotrexate and cytarabine were administered with cranial radiotherapy. The patient is currently in complete remission 12 months after the diagnosis of ALL.

Cranial nerve palsies as the first presentation of lymphoproliferative disorders, although uncommon, have already been reported.^{1,3} There is a single case report of transient third-nerve palsy in primary meningeal lymphoma.⁴ We believe that this is the first description of a patient with a systemic lymphoproliferative disorder, namely ALL, presenting with a spontaneously resolving ocular motor deficit. The cause of the left sixth-nerve palsy in our patient was most probably leukaemic involvement of the left cavernous sinus, as was shown by MRI. The transitory nature of the neurological dysfunction may be due to transient pressure by leukaemic cells on the cranial nerve itself or on the vasa nervorum, with subsequent ischaemia. The exact mechanism of transiency remains obscure, however. To conclude, cranial nerve palsy led to the diagnosis of ALL with minimal systemic clues. Spontaneous remission of a neurological deficit should not mean that an underlying malignancy can be ruled out.

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